Sarcoma of the Maxilla and the Maxillary Sinus

—Report of 7 cases—

SHIGEJIRO KURITA, KAZUTO NAGATA, SHIGENOBU MIHASHI, KATSUHIKO MIHASHI AND KOICHI MATSUO

Department of Otolaryngology, Kurume University, School of Medicine, Kurume, 830 Japan.

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Summary: Seven cases of sarcoma of the maxilla or the maxillary sinus, which were treated at the Kurume University Hospital, were reported. Histologically, 2 cases were osteosarcoma, 1 leiomyosarcoma, 1 rhabdomyosarcoma, 1 fibrosarcoma, 1 mesenchymal chondrosarcoma, and 1 chondrosarcoma. Their age ranged from 12 to 67 years with a mean age of 36 years. Curative treatments were performed in 5 cases. Of these 5 cases, 1 case of osteosarcoma died within a year and 1 case of chondrosarcoma died 2 years and a month after initial treatment. The other 3 cases, which were a leiomyosarcoma, a fibrosarcoma and a mesenchymal chondrosarcoma, are alive. Two cases, which were osteosarcoma and rhabdomyosarcoma, were inoperable. Palliative treatments were done for them, but they died within a year.

Key words: sarcoma—osteosarcoma—leiomyosarcoma—rhabdomyosarcoma—fibrosarcoma—mesenchymal chondrosarcoma—chondrosarcoma—maxillary sinus—maxilla

Introduction

Sarcomas of the maxilla and the maxillary sinus are much rarer than carcinomas of the maxillary sinus. During the 10 years from January 1, 1971 to December 31, 1980, 7 patients with histologically proven sarcoma of the maxilla or the maxillary sinus were treated at the Department of Otolaryngology, Kurume University Hospital. This paper presents a report of these cases.

Case Report

Case 1. Y. T.

A 22-year-old male noticed feeling of numbness on the right upper gingiva early in July 1972. The patient was admitted to another hospital and treated with $^{60}$Co irradiation and surgical operation. Diagnosis was sarcoma of the maxilla. No further histological information was available. He was discharged on December 13, 1972.

Early in September 1973, he developed swelling of the right upper gingiva and headache. The patient was admitted to our hospital on September 28, 1973. On physical examination, a hemorrhagic tumor of 7 cm × 7 cm in size was found on the right upper gingiva. Another subcutaneous tumor of bone-like consistency, 7 cm × 7 cm in size, was observed latero-superior to the right orbit. No cervical lymphnode was palpable. On a plain X-ray film calcification was observed in the tumor shadows. The right edge of the orbit was destroyed. On a tomodgram, the tumor associated with calcification appeared to invade the infratemporal fossa and the orbit (Fig. 1). From these
Fig. 1. An X-ray tomogram of Case 1. Dense tumor shadows (arrows) are noted in the right maxilla and frontal bone. There is an evidence of tumor invasion into the infratemporal fossa and the orbit.

X-ray findings, osteosarcoma was most suspected. This was histologically confirmed by biopsy.

Radical operation appeared to be impossible. Therefore, radiation therapy and chemotherapy were chosen as palliative treatments. A total of 6000 rad with ^{60}Co unit was irradiated and VEMP (Vincentine, Endoxan, 6-Mercaptoprine, Predonin) was applied. These therapies, however, were not effective. The patient was discharged on December 11, 1973 and referred to another hospital for palliation. He died on August 13, 1974.

Case 2. S. K.

A 55-year-old male developed epiphora and exophthalmus on the left side early in November 1974. He developed ipsilateral nasal obstruction in March, 1975. He was admitted to our hospital on March 11, 1975. Rhinoscopy revealed a mass in the left nasal cavity. X-ray films showed a mass lesion in the left nasal cavity and maxillary sinus. The bone of the inner and superior walls of the maxillary sinus was destroyed (Fig. 2). Pre-operative radiation, 3000 rad in total, accompanied by intra-arterial infusion (5Fu, BudR) into the maxillary artery via the superficial temporal artery was given to the patient. On March 20, 1975, partial maxillectomy and cryosurgery was performed. Maxillary sinus and ethmoid sinus were filled with tumor. Histological diagnosis was leiomyosarcoma. Necrotomy was done for 1 and a half months after the surgery. However, recurrence of the primary lesion was found at the inferior wall of the maxillary sinus on May 10. The recurrent tumor, including the hard palate, was removed on June 9, 1975.

The patient is currently alive 6 years and 4 months after the initial treatment without any evidence of recurrent tumor.

Case 3. M. Y.

A 12-year-old boy developed swelling of the right cheek region late in December 1975. He also complained of headache and
numbness at the right cheek region. He was admitted to our hospital on January 20, 1976. On physical examination, a subcutaneous mass, 4.5 cm × 3 cm in size, was found at the right pre-auricular region. Facial palsy was not observed. The tumor extended to the lateral and postero-superior wall of the epipharynx. Conductive deafness was noted on the right side. Tomograms revealed massive opacity in the maxillary, ethmoid and sphenoid sinuses and also a partial bone destruction of the lateral wall of the maxillary sinus (Fig. 3). Angiography suggested the existence of invasion to the cranial base. Biopsy was attempted from the mass in the epipharynx on February 2. But the tissue examined was normal mucosa of the epipharynx. Biopsy was done under maxillotomy on February 20 and histological diagnosis was rhabdomyosarcoma. The patient was palliatively treated with radiotherapy, but he became unconscious on March 8 and died on March 13, 1976.

Case 4. H. G.

A 67-year-old male noticed swelling and tenderness of the left cheek region followed by ipsilateral nasal obstruction, visual disturbance and exophthalmus in August 1977. Clinical diagnosis was carcinoma of the maxillary sinus.

On admission, a hemorrhagic bone-firm mass was observed in the left nasal cavity. No cervical lymphnode was palpable. X-ray examinations revealed a high density mass lesion in the left nasal cavity and maxillary sinus. Bone destruction was observed at the inner and superior walls of the maxillary sinus (Fig. 4).

Intra-arterial infusion of 5 Fu and BudR into the maxillary artery via the superficial temporal artery was done 5 times and total maxillectomy was performed on September 19, 1977. The maxillary sinus, ethmoid

Fig. 3. An X-ray tomogram of Case 3. There is a bone destruction (arrow) of the lateral wall of the right maxillary sinus. A massive opacity (double arrow) is observed in the maxillary, ethmoid and sphenoid sinuses.
Fig. 4. Plain X-ray films and tomogram of Case 4. High density mass lesion is noted in the left maxillary sinus and nasal cavity (arrow). There is a bone destruction of the inner and superior wall of the maxillary sinus.

sinus and nasal cavity were filled with cauliflower-like tumor. A total of 2000 rad (60Co) was given to the maxillary sinus and the nasal cavity before and after the operation. Histological examination revealed that the tumor was osteosarcoma. Necrotoomy was performed for 2 months after operation. But the recurrence of primary lesion was found at the lateral wall of the maxillary cavity on December 6. The recurrent tumor was removed on December 15.

On January 4, chest X-ray examination revealed a distant metastasis to the lung. Bloody pleural effusion was also observed. Cytological examination of the pleural effusion presented cells of class V.

The patient died one month after the detection of the distant metastasis.

Autopsy revealed distant metastasis in the lung, liver, kidney, adrenal gland, diaphragm, stomach, rectum, para-aortic lymphnodes and hilar lymphnodes.

Case 5. T.S.

A 37-year-old female was first seen in our hospital in 1963. Under the diagnosis of fibrosarcoma of the right maxillary sinus, partial maxillectomy and radiotherapy were performed. She had been doing well until October 1976, when she developed swelling of the right cheek region. Early in November, ipsilateral nasal bleeding occurred.

She came to our clinic on January 14, 1977. A hemorrhagic tumor was found in the right nasal cavity and upper gingiva. Plain X-ray films and tomograms showed a mass lesion in the right maxillary sinus and bone destruction of the lateral and inferior walls of the maxillary sinus (Fig. 5).

From January 26 to February 10, intra-arterial infusion of 5Fu and BudR and preoperative radiation (3000 rad in total) were performed. Total maxillectomy was performed on March 3, 1977. The lower aspect of the orbit was covered with a free
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Fig. 5. X-ray tomograms of Case 5. Tumor shadow (arrow) is noted in the right maxillary sinus. There is an evidence of bone destruction of the lateral and inferior walls of the maxillary sinus.

skin graft taken from the abdomen. Histological diagnosis was fibrosarcoma. After the operation, necrotomy was done for eight months. The patient was discharged on November 4, 1977. The patient is currently alive 4 years and 9 months after the operation without any evidence of recurrent tumor.

Case 6. J. A.

A 24-year-old female noticed nasal obstruction and swelling of the right cheek region late in January 1977. She visited another hospital, where histological examination presented reticulum cell sarcoma. Radiotherapy (4000 rad in total) and chemotherapy (VEMP) were done, but the tumor did not become smaller. Consequently, she was referred to a second hospital. Exploratory maxillotomy was performed for the purpose of biopsy. The histology was diagnosed as a mesenchymal chondrosarcoma. On September 7, 1977, the patient underwent mass reduction surgery which was followed by daily necrotomy. 5FU-DS (5-Fuoro-uracil dry syrup) was administered for 4 weeks after surgery (5600 mg in total). Early in June 1979, the patient developed swelling of the right infraorbital region and was referred to our hospital on July 11, 1979. Tomograms and CT scan revealed a tumor shadow at the upper wall of the right maxillary sinus (Fig. 6). Total maxillectomy and ophthalmectomy was performed on July 16, 1979. Tumor was located on the inferior wall of the orbit and invaded the orbit, being 3 cm × 2.5 cm × 2.0 cm in size. The patient is currently alive 2 years after the operation without any evidence of recurrent tumor.

Case 7. Y. O.

A 25-year-old female complained of toothache of the right upper teeth early in May 1978. The patient developed a mass of the right upper gingiva extending to the hard palate in June. The patient was admitted to our hospital on July 10.

On physical examination, a hard mass, 5 cm × 3 cm in size, was noted at the palate.
Fig. 6. An X-ray tomogram and CT scan of Case 6. Tumor shadow (arrow) is noted in the right maxillary sinus.

Fig. 7. An X-ray tomogram and CT scan of Case 7. Tumor shadow (arrow) is noted in the right maxillary sinus. Bone destruction of the medial wall of the maxillary sinus is observed.
and gingiva. Cervical lymphnodes were not palpable. Plain X-ray film showed diffuse cloudiness in the right maxillary sinus. Tomograms revealed a lobulated tumor, partially calcified, in the maxillary sinus and bone destruction of the medial and lateral walls of the maxillary sinus (Fig. 7-a). CT scan showed that the tumor invaded the posterior wall of the sinus and the infratemporal fossa (Fig. 7-b).

On July 15, 1978, biopsy was done from the tumor of the palate. The histological diagnosis was chondromyxoid fibroma. Intra-arterial infusion (5Fu, BudR) and irradiation were started after the biopsy. Since malignant tumor was most suspected clinically, biopsy was repeated. The final histological diagnosis was chondrosarcoma.

Partial maxillectomy was performed on August 10, 1978. The maxillary sinus was filled with lobulated, cartilage-hard tumor.

Two months after the operation, an evidence of recurrence was observed on the right upper gingiva. The recurrent tumor was vaporized with laser. However, the second recurrence occurred 5 months after the laser vaporization. Since the recurrent tumor invaded the orbit and infratemporal fossa, surgical removal was not possible. The patient died 25 months after the initial treatment.

Autopsy demonstrated a neoplastic invasion into the cranial base and metastasis to the lung.

Discussion

Malignant tumors of the maxillary sinus are the second frequent in head and neck malignancies in Japan. Most of them, however, are carcinoma and the incidence of sarcoma of the maxillary sinus is small. During the period of 10 years, we had only 7 cases of sarcoma while there were 124 cases of carcinoma of the maxillary sinus. The age of the sarcoma patients ranged from 12 to 67 with a mean of 36.4. In the following paragraphs, each type of sarcoma will be discussed separately.

1. Osteosarcoma

Fu and Perzin (1974), Inuyama et al. (1971), and Ohtsuki et al. (1978) reported that osteosarcoma of the maxillary sinus had poor prognosis. Our two cases of osteosarcoma died within 1 year from the initial treatment.

Osteosarcoma of the maxillary sinus should be treated with partial or total maxillectomy, since radiotherapy appears to have little effect on osteosarcoma. One of our 2 cases was inoperable and palliative radiotherapy proved to be ineffective. The other case underwent a combination therapy but the result was discouraging.

2. Leiomyosarcoma

Leiomyosarcoma of the maxillary sinus is a very rare condition of all malignant tumors in the head and neck (Inuyama et al. 1971, Fu and Perzin 1974). Fu and Perzin (1974) reported that there were only 6 cases of leiomyosarcoma in 100 non-epithelial malignant tumors of the paranasal sinuses, nasal cavity and nasopharynx. Prognosis of leiomyosarcoma has been reported to be poor (Suzuki et al. 1979, Inuyama et al. 1971). Irradiation and chemotherapy are not so effective to leiomyosarcoma. Radical operation should be indicated as the initial treatment. Our case was successfully treated with partial maxillectomy combined with irradiation and intra-arterial infusion.

3. Rhabdomyosarcoma

Rhabdomyosarcomas of the maxillary sinus, nasal cavity and nasopharynx are not common disease but more frequent than leiomyosarcomas (Fu and Perzin. 1974). Inuyama et al. (1971) reported Rhabdomyosarcoma of the maxillary sinus should be treated with surgery followed by postoperative radiation. Our case was inoperable because of an invasion into the cranial base.
Radiotherapy was done for a palliative purpose, but we could not arrest the progress of the tumor.

4. Fibrosarcoma

Fibrosarcoma of the maxillary sinus, nasal cavity and nasopharynx (Fu and Perzin, 1974) is not very rare condition. Our case was successfully treated with total maxillectomy combined with radiation and intra-arterial infusion. Radiation therapy is not so effective to fibrosarcoma (Inuyama et al. 1971). Fibrosarcoma of the maxillary sinus should be treated chiefly with surgery.

5. Mesenchymal chondrosarcoma

Mesenchymal chondrosarcoma of the maxillary sinus is very rare. In Japan, only 3 cases of mesenchymal chondrosarcoma of the maxillary sinus have been reported. The present case is 1 of these 3 cases and was previously reported by Minami et al. (1979). This pathology should be treated surgically since radiotherapy and chemotherapy are not effective.

6. Chondrosarcoma

Chondrosarcoma of the maxilla is rare and its prognosis is poor. Arlen et al. (1970) reported that the 5-year survival rate of chondrosarcoma of the maxilla was 40% following surgical treatments. Irradiation is not so effective to chondrosarcoma. Chondrosarcoma of the maxilla should be treated with complete extirpation (Inuyama et al. 1971). Prognosis depends on the success of the initial operation.

Our case was treated with partial maxillectomy combined with intra-arterial infusion and irradiation, but a local recurrence occured 2 months later. The recurrence was not controlled and the patient died 2 years and 1 month after the initial treatment. The extent of removal at the initial treatment should have been insufficient.

References


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